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Causes of Pulmonary Arterial Hypertension in Down's Syndrome

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Children with Down syndrome (DS) have an increased risk for developing pulmonary arterial hypertension (PAH) due to multiple factors: Congenital heart disease with persistent left-to-right shunts, chronic upper airway obstruction, abnormal pulmonary vasculature growth, alveolar hypoventilation and recurrent pulmonary infection.

Congenital cardiac defects are reported in 19-43% of Down's syndrome. Common lesion is an endocardial cushion defect in 43%. DS and CHD seem to develop PAH at a faster rate and have persistent disease after cardiac surgery compared to non-DS patients with similar defects.

Upper airway obstruction is common in DS due to: midfacial hypoplasia, macroglossia, narrowing of the nasopharynx, tonsillar and adenoidal enlargement, laryngomalacia, tracheomalacia and congenital malformations of the larynx and the trachea. The incidence of OSA was reported to be 30-50%. Exacerbating factors including obesity and gastro-esophageal reflux may contribute to the occurrence of sleep apnea.

The aim of this paper is to review the causes of pulmonary hypertension in DS population and its management.